A case of pulmonary arteriovenous malformation (PAVM) in a 32-year-old woman who underwent successful left upper lobectomy is presented. A whole-body technetium-99m-labeled macro-aggregated albumin scan was used to demonstrate intrapulmonary right-to-left shunt, and shunt fraction was calculated as 39%. The patient underwent a follow-up scan after the operation; shunt fraction was calculated within normal limits (5%). Her follow-up visit at the third month showed no cyanosis and no dyspnea on exertion, and she had an arterial blood oxygen saturation of 97% at room air.

Pulmonary arteriovenous malformations (PAVMs) are caused by abnormal communications between pulmonary arteries and pulmonary veins, which are most commonly congenital in nature. These lesions are quite uncommon. In an autopsy study in 1953 from Johns Hopkins Hospital, only 3 cases of PAVM were detected in 15,000 consecutive autopsies [1].

A 32-year-old woman presented with a 31-year history of cyanosis. The patient had a history of right temporal brain abscess drainage at 22 years of age. She had no family history of hereditary hemorrhagic telangiectasia. A laboratory database revealed low arterial blood oxygen saturation (83%) and polycythemia with cyanosis. Her hemoglobin value was 20.4 g/dL. On physical examination, she had clubbing of her fingers and an upper lobe chest bruit over the posterior left hemithorax, which was increasing in pitch and volume with inspiration.

A spiral chest computed tomographic scan with intravenous contrast revealed two high-density large nodular lesions located in the apical and lingular segments of the upper lobe.

A whole-body scan was performed with 10,000 technetium-99m-labeled macro-aggregated albumin particles to rule out a right-to-left shunt. It showed markedly extrapulmonary tracer distribution in both kidneys and in the brain (Fig 1). The shunt fraction was calculated as (39%) using the Gates’ semiquantitative calculation method according to the following formula [2]:

\[
\text{\% right-to-left shunt} = \left(\frac{(\text{total body counts} - \text{total lung counts})}{\text{total body counts}}\right) \times 100
\]

Pulmonary angiography revealed two PAVMs in the left upper lobe with the apical one being larger than 2 cm in diameter and the lingular one being centrally localized (Fig 2).

A left upper lobectomy was performed without complication. Histolopathologic study of the resected lobe revealed two plexiform masses of dilated vessels with feeding vessels. Of them, the one in the apical segment measured 3 × 3 cm, and the other one in the lingular segment measured 2 × 1.5 cm.

The patient underwent a follow-up scan (Fig 1) after operation and shunt fraction was calculated within normal limits (5%). Her follow-up visit at the third month showed no cyanosis and no dyspnea on exertion, and she had an arterial blood oxygen saturation of 97% at room air.

Comment

Pulmonary arteriovenous malformation is a hereditary disease; approximately 70% of patients have hereditary hemorrhagic telangiectasia [3]. Conversely, 15% to 35% of the patients with hereditary hemorrhagic telangiectasia have PAVMs [3]. Pulmonary arteriovenous malformation occurs twice as often in females as in males, but there is a male predominance in newborns [4].

References


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Quantitative Shunt Imaging in the Evaluation of Therapeutic Surgery in a Patient With Pulmonary Arteriovenous Malformation

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A case of pulmonary arteriovenous malformation (PAVM) in a 32-year-old woman who underwent successful left upper lobectomy is presented. A whole-body technetium-99m-labeled macro-aggregated albumin scan was used to demonstrate intrapulmonary right-to-left shunt, and shunt fraction was calculated as 39%. The patient underwent a follow-up scan after the operation; shunt fraction was calculated to be within normal limits (5%).

The whole-body technetium-99m-labeled macro-aggregated albumin scan seems to be a simple and noninvasive test in the demonstration and quantitation of the degree of the right-to-left shunt in pulmonary arteriovenous malformation. The whole-body technetium-99m-labeled macro-aggregated albumin scan may also be useful for evaluation of the hemodynamic changes after surgical treatment and follow-up of the patients with pulmonary arteriovenous malformation.


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The clinical pattern of the disease is variable. The classical triad of cyanosis, polycythemia, and clubbing has been noted in approximately 20% of patients. Other frequent extrapulmonary symptoms and signs are headache in 43%, transient ischemic attacks in 57%, and cerebrovascular accidents in 18%. Pulmonary arteriovenous malformation lesions of less than 2 cm in diameter are usually asymptomatic [3]. Patients with multiple PAVM usually have two to eight lesions [3]. The classic roentgenographic appearance of a PAVM is that of a round or oval mass of uniform density, frequently lobulated but sharply defined, more commonly in the lower lobes, ranging from 1 to 5 cm in diameter [3]. A plain tomographic scan of the chest may be more accurate than plain chest roentgenography, and the presence of a PAVM and its vascular anatomy can also be evaluated by contrast-enhanced ultrafast computed tomography and three-dimensional helical computed tomography. Magnetic resonance imaging may be useful in differentiating PAVM from various types of pulmonary nodules [3]. Contrast echocardiography is an excellent tool for evaluation of intrapulmonary shunts and is able to identify small right-to-left shunts, even when they are not suggested by gas exchange data [3]. However, contrast echocardiography is somewhat limited as a first-line screening test for PAVM owing to cost, availability, and over-detection of clinically unimportant PAVM [3].

Contrast pulmonary angiography is the gold standard in the diagnosis of PAVM, and it is usually necessary if resectional or obliterator therapy is being considered [3]. In recent years, digital subtraction angiography seems to be replacing conventional angiography in the diagnosis and treatment of PAVM.

Technetium-99m-labeled macro-aggregated albumin is the most widely used agent for pulmonary perfusion scintigraphy. In normal subjects, after an antecubital vein injection, macro-aggregated albumin particles (10 to 60 μm) are microembolized in the pulmonary arterioles and pre-capillaries in accordance with pulmonary arterial blood flow, and only 2% to 5.7% of these agents shunts to the systemic circulation [5]. Thus, activity in other organs is rarely seen on lung perfusion scan, and the size of the shunt is directly proportional to the number of macro-aggregated albumin particles that reach the systemic

Fig 1. Preoperative (PREOP) whole-body 99mTc-MAA scan images showed markedly extrapulmonary tracer distribution in both kidneys (1A, 1C; arrows) and brain (1A, 1B; arrowheads). Note that postoperative (POSTOP) follow-up whole-body scan image did not demonstrate extrapulmonary uptake (2).

Fig 2. Pulmonary angiography showing two pulmonary arteriovenous malformations located in the left upper lobe.
circulation to lodge in the end-organ capillary beds, such as those in the brain, kidneys, and spleen.

The fraction of cardiac output that shunts from right to left (shunt fraction, normal ≤ 5%) is elevated in 88% to 100% of selected patients with PAVM [3]. The right-to-left shunting induces chronic hypoxemia or paradoxical embolism. The exact shunt fraction can be calculated with either radionuclide method (Gates’ method) or 100% oxygen method. The 100% oxygen method involves measurement of PaO2 and SaO2 after breathing 100% oxygen for 15 to 20 minutes [6]. A shunt fraction of greater than 5% by this method is considered abnormal and warrants additional workup.

The radionuclide method of shunt calculation has several advantages (ie, simple, noninvasive, nonoperator dependent) in comparison with the 100% oxygen method (ie, arterial blood sampling is not needed and the 100% oxygen method may overestimate the intrapulmonary shunt). However, the radionuclide method is not routinely available in some hospitals [3].

An alternative approach is to measure SaO2 and PaO2 while breathing room air. PaO2 > 90 mm Hg and SaO2 > 96.5% effectively rule out a significant shunt, whereas PaO2 < 85 mm Hg or SaO2 < 96% indicate a potential shunt fraction of > 5%. This method is less specific than the 100% oxygen method because it does not differentiate shunt from hypoxemia due to ventilation-perfusion mismatch [3].

It is recommended that all symptomatic PAVM and PAVM > 2 cm in diameter be treated with either surgery or embolotherapy [3].

In conclusion, whole-body technetium-99m-labeled macro-aggregated albumin radionuclide imaging seems to be a simple, noninvasive, nonoperator-dependent method that is useful in demonstration and quantitation of the degree of the right-to-left shunt in PAVM. The whole-body technetium-99m-labeled macro-aggregated albumin scan may also be useful for evaluation of the hemodynamic changes after surgical treatment and follow-up of patients with PAVM.

References


Bilateral Intrathoracic Kidneys and Adrenal Glands Associated With Posterior Congenital Diaphragmatic Hernias

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We present a case of bilateral intrathoracic kidneys and adrenal glands associated with bilateral posterior diaphragmatic defects in a symptomatic 18-month-old baby boy. The diaphragmatic defect did not appear to be the typical posterolateral diaphragmatic hernia of Bochdalek. The patient underwent primary surgical correction through an abdominal approach. Postoperatively, the patient enjoyed an uneventful course and was discharged home without any further events. We discuss this report of bilateral intrathoracic kidneys associated with bilateral diaphragmatic hernias, we describe the operative management, and we analyze the possible embryological development of this defect.


Ectopic kidneys are not an uncommon phenomena occurring in 1% of postmortem examinations, but complete intrathoracic cases are very rare representing only 5% of all renal ectopia. These are usually asymptomatic in males (2:1), located on the left side (1.5:1), and are diagnosed in all ages [1, 2]. Only about 150 cases of thoracic kidneys have been reported in the last century, and of these only four have been bilateral [1]. No other case of complete bilateral intrathoracic kidneys and adrenal glands with congenital diaphragmatic hernias (CDHs) was found in the literature. We present the case of an 18-month-old baby boy with bilateral intrathoracic kidneys and adrenal glands associated with posterior diaphragmatic defects that did not seem to be consistent with the classic Bochdalek diaphragmatic hernia.

The patient is an 18-month-old Hispanic, male baby who was delivered at full term to a G1P1 mother by cesarean section. He was treated for recurrent tonsillitis and upper respiratory tract infections with a tonsillectomy and adenoidectomy. After his surgery, there was substantial